General

Guideline Title

British Thoracic Society guideline for respiratory management of children with neuromuscular weakness.

Bibliographic Source(s)

Hull J, Aniapravan R, Chan E, Chatwin M, Forton J, Gallagher J, Gibson N, Gordon J, Hughes I, McCulloch R, Russell RR, Simonds A. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. Thorax. 2012 Jul;67(Suppl 1):i1-i40. [270 references] PubMed

Guideline Status

This is the current release of the guideline.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- March 22, 2016 – Opioid pain medicines : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

Recommendations

Major Recommendations

The levels of evidence (1++, 1+, 1-, 2++, 2+, 2-, 3, 4), grades of recommendations (A-D), and good practice points (GPPs) are defined at the end of the "Major Recommendations" field.

Summary of Recommendations

Identifying Children at Risk of Respiratory Complications

- Clinical assessment of respiratory health should be part of every medical consultation for children with neuromuscular weakness (NMW) and should be directed towards identifying progressive muscle weakness, ability to cope with respiratory infection, aspiration, progression of scoliosis and sleep-disordered breathing. [D]
- Ulna length or arm span should be used to predict lung function in children with neuromuscular disease whose height cannot be accurately measured. [B]
- Vital capacity should be measured in all patients with neuromuscular disease who are capable of performing spirometry as part of the respiratory assessment. [C]
- Cough peak flow should be used as part of the assessment of effective secretion clearance in children with neuromuscular disease over the age of 12 years. [GPP]
- Assessment for sleep-disordered breathing should be carried out no less than annually for children with neuromuscular disease who have a vital capacity of <60% predicted and for children who have become non-ambulant because of progressive muscle weakness or who never attain the ability to walk. [D]
- Assessment for sleep-disordered breathing should be carried out no less than annually for all infants with weakness, children with NMW who have symptoms of obstructive sleep apnoea or hypoventilation, children with clinically apparent diaphragmatic weakness and children with rigid spine syndromes. [GPP]
- In young children whose rate of disease progression is uncertain, or in older children who have shown a clinical deterioration or who are suffering with repeated infections, or who develop symptoms of sleep-disordered breathing, sleep assessment may need to be more frequent than once a year. [GPP]
- All children with abnormal overnight oximetry should undergo more detailed sleep monitoring with at least oxycapnography. [GPP]
- When there is doubt about the cause of sleep disordered breathing, overnight polysomnography or sleep polygraphy should be performed. [GPP]
- Portable overnight oxycapnography or polygraphy in the home may be the most appropriate option for some patients. [GPP]
- Children with neuromuscular disease with a history of swallowing difficulties should have a feeding assessment by a speech and language therapist including a video fluoroscopy swallow assessment if the swallow is thought to be unsafe. [GPP]

Airway Clearance and Respiratory Muscle Training

- Children with ineffective cough (including children over 12 years of age with cough peak flow <270 litres/min), particularly if they have had episodes of deterioration with respiratory infection, should be taught augmented cough techniques. [C]
- Manual cough assist and air-stacking methods to achieve maximum insufflation capacity are effective methods of improving cough efficiency and should be used when appropriate. [C]
- Mechanical insufflation/exsufflation (MI-E) should be considered in very weak children, those with loss of bulbar function, and those who cannot cooperate with manual cough assist or air-stacking or in whom these methods are not effective. [C]
- Oscillatory techniques such as high-frequency chest wall oscillation and intrapulmonary percussive ventilation should be considered in children who have difficulty mobilising secretions or who have persistent atelectasis, despite use of other airway clearance techniques. [D]
- Airway clearance techniques should be used during respiratory infection when oxygen saturation levels fall below 95% while the child is breathing room air. If the techniques being used fail to result in an increase in oxygen saturation to 95% or above, different methods of airway clearance should be used. This may require attendance at hospital for treatment. [D]
- MI-E should be available in the acute setting in all hospitals that treat neuromuscular patients as an alternative method of airway clearance with the purpose of preventing deterioration and the need for intubation and mechanical ventilation. [D]
- Nebulised normal saline may be considered in children who have continued tenacious secretions. [GPP]
- Humidification should be considered in children who use non-invasive ventilation (NIV) and who have tenacious airway secretions. Care should be taken to ensure that use of humidification does not result in a troublesome increase in oral secretions. [GPP]
- When using sputum mobilising techniques, appropriate emergency equipment (e.g., resuscitation bag and suction) should be available in case of mobilising large mucus plugs into the central airways where they may result in airways obstruction. [GPP]
- Children who use regular night-time or diurnal NIV should use their ventilator to support deep breathing during airway clearance treatments. Use of NIV during airway clearance sessions can help prevent respiratory muscle fatigue. [GPP]
- Children who use MI-E for airway clearance should be given long enough periods of rest during treatment sessions to prevent respiratory muscle fatigue due to coughing. [GPP]
- At the end of a treatment session with MI-E it is important to complete the session with an insufflation to leave the child with an appropriate functional residual capacity. [GPP]

Assisted Ventilation

- Children with NMW resulting in symptomatic nocturnal hypoventilation or daytime hypercapnia should be supported with NIV. [C]

- A non-invasive approach should be considered in children needing daytime ventilation. [D]
- Clinical teams caring for children using home ventilators should become familiar with a small number of machines. For most children pressure-targeted machines work well and are simple to use. [GPP]
- Ventilation modes with fixed inspiratory times are usually the most appropriate for use in young or very weak children. [GPP]
- For older teenage children who need to use mouthpiece ventilation during the day, and/or who use air-stacking as a way of assisting cough, volume-targeted or hybrid ventilators may be preferred. [GPP]
- When full face masks are used, anti-asphyxia valves should be fitted to allow room air breathing in case of ventilator failure. The risks of the child vomiting and aspirating should be considered, particularly if the child is unable to remove the mask on their own. [GPP]
- Children needing ventilatory support for more than 16 hours per day should be provided with two ventilators in case of equipment failure. [GPP]
- Once a child has become established on NIV, a sleep polygraphy or oxycapnography should be carried out to check that it has effectively abolished sleep-associated hypoventilation. Ventilator settings should be adjusted and rechecked as necessary. [GPP]
- Children supported by NIV should be assessed regularly with repeat sleep studies to ensure continued effectiveness of NIV at preventing hypoventilation. The frequency of review will vary according to clinical circumstances, but should not be less than every 12 months. [D]
- Assessment for skin injury and facial flattening should be carried out regularly in children using NIV and the mask interface adjusted as necessary to minimise these complications. [GPP]
- Tracheostomy tubes should be carefully sized and sited to ensure the tip of the tube does not abut the tracheal wall. [GPP]
- Family and child preference should be taken into account when considering tracheostomy to facilitate diurnal ventilation. [GPP]
- Oxygen alone should not be used to correct hypoxaemia caused by hypoventilation in patients with neuromuscular disease. [GPP]
- NIV should be the first-line treatment for children with NMW in acute respiratory failure. [D]
- Intensive care units caring for children with NMW should be aware of appropriate extubation criteria. These should include the presence of only minimal airway secretions, use of effective airway clearance methods (such as MI-E devices) and oxygen saturation more than 94% without supplemental oxygen for more than 12 hours. Continuous NIV should be used immediately after extubation. [GPP]

Planning for Surgical Procedures

- Surgery in children with NMW should take place in units with experienced paediatric surgeons, anaesthetists and physiotherapists, and where there are facilities for paediatric intensive care and NIV. [D]
- Extubation protocols for high-risk children (those with vital capacity <60% predicted and/or ineffective cough and/or already use NIV for hypoventilation) should include effective airway clearance techniques and immediate use of NIV following extubation. [GPP]
- Children with NMW who require surgery (including scoliosis surgery) should be assessed by a multidisciplinary team prior to any intervention. [GPP]

Scoliosis

- The effect of wearing a spinal brace on respiratory function should be assessed and weighed against the limited evidence of benefit in terms of affecting final scoliosis severity. [D]
- The primary consideration when planning surgery for children with scoliosis associated with NMW should be comfort and quality of life. [GPP]

Other Problems and Interventions That Impact on Respiratory Health

- Videofluoroscopy and feeding advice from a specialist therapist is indicated in children with a history of recurrent chest infection or swallowing difficulties. [GPP]
- A problem-orientated approach to nutrition should aim to minimise risk of aspiration, optimise nutritional status, promote comfort and balance the positive social consequences of continued oral feeding. [GPP]

Transition to Adult Care

- A key worker can act as a valuable advocate, a source of knowledge and provide support for young people with neuromuscular disease and their families during the period of transition from paediatric to adult services. [GPP]

Quality of Life and Palliative Care

- Evaluating the child and parent or carer using multidimensional health-related quality of life assessment tools should be the standard for routine assessment in clinical practice and future clinical trials. [D]
- International standardised disease-specific tools for children and young people with neuromuscular disorders should be used if available to

evaluate clinical interventions and patient- related outcome measures with respect to quality of life. [D]

- Assisting patients, parents and carers to make informed choices that are consistent with their own values and preferences requires physicians to engage patients and their parents and carers in a process of mutual participation in decision-making with full disclosure of all information in a sensitive and timely fashion. [D]
- Open discussion across the multidisciplinary team regarding the type and duration of specific interventions encourages transparency and shared decision-making. [GPP]
- Benefit and burden of all interventions must be considered with respect to impact on mental health of patients and their families. [D]
- Acknowledgement of stress factors and mental health issues, requirement for wider support through organizations, and practical and focused individual care are needed for patients and their parents or carers. [D]
- Routine pain evaluation should be part of standard clinical assessment in all children and young people with neuromuscular disorders. [D]
- Attention to the wider physical impact of chronic illness through vigilant symptom management is required. [D]
- Dyspnoea is a subjective feeling, which can respond to non-drug measures and treatment directed at the cause. [GPP]
- Low-dose opioids may be used to manage distressing breathlessness even when active treatment is pursued. With slow titration of opioids, respiratory depression is extremely unlikely. [GPP]
- In the terminal phase of respiratory failure, symptoms must be reassessed frequently and can be effectively managed by a multimodal approach, including careful titration of opioids and psychotropic agents. [GPP]
- Advance care planning should be an integral part of the active management of children and young people with neuromuscular disorders. Advance care plans can be used as a vehicle for information exchange and considered decision-making. [D]
- Patients and families need to have ownership of the advance care plan and be educated as to its uses. [D]
- Advance care plans should be reviewed by the multidisciplinary team on a regular basis. [GPP]
- Families need access to skilled experts for multidimensional coordinated palliative care support, providing regular review of their needs at various stages in their condition. [D]
- Generic palliative care skills should be cascaded to other professionals providing neuromuscular services. [GPP]

Special Considerations

- Carers and parents of children with severe NMW, including all those using NIV and all those with tracheostomy, should have basic life support training. [GPP]
- Written plans for the management of acute exacerbations, which include details of effective airway clearance methods and ventilator settings when appropriate, and contact details of relevant healthcare professionals are recommended. [GPP]

Definitions:

Revised Grading System for Recommendations in Evidence-based Guidelines

Grade	Evidence
1++	High-quality meta-analyses, systematic reviews of randomised controlled trials (RCTs) or RCTs with a very low risk of bias
1+	Well conducted meta-analyses, systematic reviews of RCTs or RCTs with a low risk of bias
1-	Meta-analyses, systematic reviews or RCTs or RCTs with a high risk of bias
2++	High-quality systematic reviews of case-control or cohort studies or high-quality case-control or cohort studies with a very low risk of confounding, bias or chance, and a high probability that the relationship is causal
2+	Well conducted case-control or cohort studies with a low risk of confounding, bias or chance, and a moderate probability that the relationship is causal
2-	Case-control or cohort studies with a high risk of confounding, bias or chance and a significant risk that the relationship is not causal
3	Non-analytic studies, for example, case reports, case series
4	Expert opinion

Grades of Recommendation

Grade	Type of Evidence

Grade	Type of Evidence
A	At least one meta-analysis, systematic review, or randomised controlled trial (RCT) rated as 1++ and directly applicable to the target population *or* A systematic review of RCTs or a body of evidence consisting principally of studies rated as 1+ directly applicable to the target
B	A body of evidence including studies rated as 2++ directly applicable to the target population and demonstrating overall consistency of results *or* Extrapolated evidence from studies rated as 1++ or 1+
C	A body of evidence including studies rated as 2+ directly applicable to the target population and demonstrating overall consistency of results *or* Extrapolated evidence from studies rated as 2++
D	Evidence level 3 or 4 *or* Extrapolated evidence from studies rated as 2+
GPP (Good Practice Point)	Important practical points for which there is no research evidence, nor is there likely to be any research evidence. The Guideline Committee wishes to emphasise these as good practice points

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Neuromuscular conditions, including: Spinal muscular atrophy (SMA), SMA with respiratory distress, Duchenne/Becker muscular dystrophy, limb girdle muscular dystrophy, facioscapulohumeral muscular dystrophy, Emery-Dreifuss muscular dystrophy, congenital muscular dystrophies, rigid spine syndrome and rigid spine muscular dystrophy, congenital myopathies, myotonic dystrophy, congenital myasthenic syndromes, mitochondrial myopathies, Charcot-Marie-Tooth disease, Pompe disease

Guideline Category

Evaluation

Management

Risk Assessment

Treatment

Clinical Specialty

Anesthesiology

Critical Care

Family Practice

Neurology

Orthopedic Surgery

Pediatrics

Physical Medicine and Rehabilitation

Pulmonary Medicine

Sleep Medicine

Speech-Language Pathology

Intended Users

Advanced Practice Nurses

Allied Health Personnel

Nurses

Physical Therapists

Physician Assistants

Physicians

Respiratory Care Practitioners

Speech-Language Pathologists

Guideline Objective(s)

To summarise the available evidence in the field of respiratory management of children with neuromuscular weakness and to provide recommendations that will aid the healthcare professional in delivering good quality patient care

Target Population

Children with neuromuscular weakness and respiratory problems

Note: The following areas fall outside the scope of the guideline:

Non-respiratory aspects of management of children with neuromuscular disease
Detailed anaesthetic management of children with neuromuscular disease
Adults with neuromuscular disease
Children with cerebral palsy
Children with myasthenia gravis
Children with Guillain-Barré syndrome

Interventions and Practices Considered

Risk Assessment/Evaluation

1. Ulna length or arm span measurement (in children whose height cannot be accurately measured)
2. Vital capacity measured
3. Cough peak flow

4. Assessment for sleep-disordered breathing
5. Sleep polygraphy or oxycapnography
6. Feeding assessment
7. Assessment of wearing a spine brace on respiratory function in patients with scoliosis
8. Assessment for skin injury and facial flattening should be carried out regularly in children using non-invasive ventilation (NIV)
9. Multidimensional health-related quality of life assessment tools
10. International standardised disease-specific tools to evaluate clinical interventions
11. Routine pain evaluation

Management/Treatment

1. Airway clearance and respiratory muscle training
    - Augmented cough techniques
    - Manual cough assist and air-stacking methods
    - Mechanical insufflation/exsufflation (MI-E)
    - Oscillatory techniques
    - Nebulised normal saline
    - Humidification
    - Sputum mobilizing techniques (with appropriate emergency equipment e.g., resuscitation bag and suction)
2. Assisted ventilation
    - NIV (with humidification)
    - Volume targeted, hybrid ventilators
3. Planning for surgical procedures
    - Performance of surgery in units with experienced paediatric surgeons, anaesthetists and physiotherapists, and where there are facilities for paediatric intensive care and NIV
    - Extubation protocols
    - Multidisciplinary team assessment
4. Videofluoroscopy and feeding advice
5. Nutritional advice
6. Management of transition to adult care
7. Quality of life and palliative care
    - Acknowledgement of stress factors and mental health issues,
    - Open discussion across the multidisciplinary team regarding the type and duration of specific interventions, transparency and shared decision-making
    - Consideration of benefit and burden of all interventions with respect to impact on mental health of patients and their families
    - Symptom management
    - Low-dose opioids
    - Management of terminal phase of respiratory failure
    - Advance care plans
8. Life support training for carers and parents of children with severe NMW
9. Written plans for the management of acute exacerbations

Major Outcomes Considered

- Sensitivity and specificity and predictive value of respiratory assessments
- Quality of life
- Respiratory symptoms
- Respiratory complications
- Progression to respiratory failure
- Hospital admission
- Adverse effects of noninvasive ventilation
- Adverse effects of tracheostomy

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Clinical Questions and Literature Search

Systematic electronic database searches were conducted to identify potentially relevant studies for inclusion in the guideline. For each topic area the following databases were searched: Ovid MEDLINE (including MEDLINE In Process), Ovid, EMBASE, EMSCO CINAHL, Ovid PsycINFO and the Cochrane Library (including the Cochrane Database of Systematic Reviews, the Database of Abstracts of Reviews of Effects, and the Cochrane Central Register of Controlled Trials).

The searches were first run in March 2010 and were updated in May 2011. Searches included a combination of indexed terms and free text terms. Searches were not limited to studies on children. The search strategy is available in online appendix S2 (see the "Availability of Companion Documents" field).

Appraisal of the Literature

Appraisal was performed using the criteria stipulated by the Appraisal of Guidelines for Research and Evaluation (AGREE) collaboration. Each paper was appraised by a pair of reviewers. One individual read the title and abstract of each article retrieved by the literature searches and decided whether the paper was definitely relevant, possibly relevant or not relevant to the project. The following criteria were formulated for categorising the abstracts into these three groups were:

- Whether the study addressed the clinical question.
- Whether the appropriate study type was used to produce the best evidence to answer the clinical question.
- The abstract was in English.
- Abstracts were not rejected on the basis of the journal of publication, the country in which the research was performed or published, or the date of publication.
- Abstracts were not rejected if they dealt only with adults provided that the topic was relevant to children with neuromuscular weakness (NMW).

The full paper was obtained for all relevant or possibly relevant abstracts and allocated to the relevant section(s). The first screening process identified 686 of the 4683 reference abstracts to be definitely or possibly relevant to the guideline. Two guideline reviewers independently reviewed the abstracts to identify papers to be appraised for the guideline.

Number of Source Documents

A total of 177 papers were critically appraised.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Revised Grading System for Recommendations in Evidence-based Guidelines

Grade	Evidence
1++	High-quality meta-analyses, systematic reviews of randomised controlled trials (RCTs) or RCTs with a very low risk of bias

Grade	Evidence
1++	Well conducted meta-analyses, systematic reviews of RCTs or RCTs with a low risk of bias
1-	Meta-analyses, systematic reviews or RCTs or RCTs with a high risk of bias
2++	High-quality systematic reviews of case-control or cohort studies or high-quality case-control or cohort studies with a very low risk of confounding, bias or chance, and a high probability that the relationship is causal
2+	Well conducted case-control or cohort studies with a low risk of confounding, bias or chance, and a moderate probability that the relationship is causal
2-	Case-control or cohort studies with a high risk of confounding, bias or chance and a significant risk that the relationship is not causal
3	Non-analytic studies, for example, case reports, case series
4	Expert opinion

Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

This guideline is based on the best available evidence. The methodology used to write the guideline adheres strictly to the criteria set out by the Appraisal of Guidelines for Research and Evaluation (AGREE) collaboration (available online at http://www.agreetrust.org/).

Appraisal of the Literature

The two leads for each section independently appraised each paper assigned to them using the Scottish Intercollegiate Guidelines Network (SIGN) critical appraisal checklists. An evidence grade was assigned to each study using the SIGN grading system (see the "Rating Scheme for the Strength of the Evidence" field) and is shown in the evidence tables (online appendix S1 see the "Availability of Companion Documents" field).

Considered Judgement and Grading of the Evidence

The Guideline Committee used evidence tables to judge the body of evidence and grade recommendations for this guideline. Evidence tables are available in online appendix S1 (see the "Availability of Companion Documents" field). When evidence was lacking to answer the formulated clinical questions, expert opinions were obtained for formal consensus statements using the Delphi method.

Methods Used to Formulate the Recommendations

Expert Consensus

Expert Consensus (Delphi)

Description of Methods Used to Formulate the Recommendations

The Guideline Committee corresponded regularly by email and meetings of the full group were held in March 2010, November 2010, March 2011 and May 2011.

The following factors were considered when grading the recommendations:

- The available volume of the body evidence.
- How applicable the obtained evidence was in making recommendations for the defined target audience of this guideline.
- Whether the evidence was generalisable to the target population for the guideline.
- Whether there was a clear consistency in the evidence obtained to support recommendations.
- What the implications of recommendations will be on clinical practice in terms of recourses and skilled expertise.
- Cost effectiveness was not reviewed in detail because in-depth economic analysis of the recommendations falls beyond the scope of this

guideline.

Recommendations were graded from A to D as indicated by the strength of the evidence (see the "Rating Scheme for the Strength of the Recommendations" field). Important practical points lacking any research evidence were highlighted as 'Good practice points' (GPP).

Rating Scheme for the Strength of the Recommendations

Grades of Recommendation

Grade	Type of Evidence
A	At least one meta-analysis, systematic review, or randomised controlled trial (RCT) rated as 1++ and directly applicable to the target population *or* A systematic review of RCTs or a body of evidence consisting principally of studies rated as 1+ directly applicable to the target
B	A body of evidence including studies rated as 2++ directly applicable to the target population and demonstrating overall consistency of results *or* Extrapolated evidence from studies rated as 1++ or 1+
C	A body of evidence including studies rated as 2+ directly applicable to the target population and demonstrating overall consistency of results *or* Extrapolated evidence from studies rated as 2++
D	Evidence level 3 or 4 *or* Extrapolated evidence from studies rated as 2+
GPP (Good Practice Point)	Important practical points for which there is no research evidence, nor is there likely to be any research evidence. The Guideline Committee wishes to emphasise these as good practice points

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

A draft guideline document was circulated to all the relevant stakeholders (see Appendix 3 in the original guideline document) for consultation in October/November 2011 and the draft guideline was made available for public consultation via the British Thoracic Society (BTS) website in November 2011. The BTS Standards of Care Committee reviewed the draft guideline in November 2011. The guideline was revised, taking account of comments from the Committee, stakeholders and feedback from public consultation, and submitted for publication in March 2012.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate management of respiratory complications in children with neuromuscular weakness

Potential Harms

- Standard chest physiotherapy: Care needs to be applied with these techniques in infants when the chest wall is very compliant and the closing volume of the lung is high, as this can induce atelectasis.
- With intrapulmonary percussive ventilation (IPV) and high-frequency chest wall oscillation (HFCWO) there is the potential to mobilise a large volume of secretions and therefore it is important to ensure that the appropriate emergency equipment (e.g., resuscitation bag and suction) is available in case of mobilising a large mucus plug into a central airway.
- Although pneumothorax is a potential risk from any positive pressure device, there have been no reports of pneumothorax directly attributable to mechanical insufflation/exsufflation (MI-E) devices in children.
- Ventilators: When full face masks are used, anti-asphyxia valves should be fitted to allow room air breathing in case of ventilator failure. The risks of the child vomiting and aspirating should be considered, particularly if the child is unable to remove the mask on their own.
- Adverse effects of non-invasive ventilation (NIV):
    - Break down of the skin, usually on the bridge of the nose or forehead, can occur when using nasal or face-mask ventilation.
    - Pressure from the nasal or face mask used for NIV on the growing face can result in under-development of the maxilla, leading to mid-face flattening and mal-occlusion of the teeth.
- Adverse effects of tracheostomies include increased respiratory secretions and respiratory infections, dysphagia, granuloma formation and tracheo-arterial fistulas with catastrophic hemorrhage.
- Children with neuromuscular weakness (NMW) are vulnerable to the adverse effects of general anaesthesia and surgery and have increased intra-operative and postoperative morbidity. Respiratory muscle strength is diminished in the postoperative period, most often due to the combined effects of pain and sedation, and can result in hypoventilation and airway secretion retention.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

End of Life Care

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Hull J, Aniapravan R, Chan E, Chatwin M, Forton J, Gallagher J, Gibson N, Gordon J, Hughes I, McCulloch R, Russell RR, Simonds A. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. Thorax. 2012 Jul;67(Suppl 1):i1-i40. [270 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2012 Jul

Guideline Developer(s)

British Thoracic Society - Medical Specialty Society

Source(s) of Funding

British Thoracic Society

Guideline Committee

Guideline Committee

Composition of Group That Authored the Guideline

Committee Members: Jeremy Hull, Department of Paediatrics, Oxford University Hospitals NHS Trust, Oxford, UK; Roona Aniapravan, Department of Paediatrics, Cambridge University Hospitals NHS Foundation Trust, Cambridge, UK; Elaine Chan, Royal Manchester Children's Hospital, Manchester, UK; Michelle Chatwin, National Institute of Health Research Respiratory Biomedical Research Unit, Royal Brompton and Harefield NHS Foundation Trust, UK; Julian Forton, University Hospital of Wales, Cardiff, UK; Jayne Gallagher, Department of Paediatrics, Oxford University Hospitals NHS Trust, Oxford, UK; Neil Gibson, Royal Hospital for Sick Children, Glasgow, UK; Jill Gordon, Child Development Centre, Suffolk Community Healthcare, UK; Imelda Hughes, Royal Manchester Children's Hospital, Manchester, UK; Renee McCulloch, Great Ormond St Hospital, London, UK; Robert Ross Russell, Department of Paediatrics, Cambridge University Hospitals, NHS Foundation Trust, Cambridge, UK; Anita Simonds, National Institute of Health Research Respiratory Biomedical Research Unit, Royal Brompton and Harefield NHS Foundation Trust, UK

Financial Disclosures/Conflicts of Interest

Contributor	Conflict of Interest
Jeremy Hull Consultant in paediatric respiratory medicine, Children's Hospital, Oxford	None
Roona Aniapravan Specialist registrar, paediatric respiratory medicine, Addenbrooke's Hospital, Cambridge	None
Elaine Chan Consultant in paediatric respiratory medicine, Royal Manchester Children's Hospital	None
Michelle Chatwin Consultant physiotherapist in respiratory support, Royal Brompton Hospital, London	None
Julian Forton Consultant in paediatric respiratory medicine, University Hospital of Wales, Cardiff	None
Jayne Gallagher Clinical nurse specialist, Children's Hospital, Oxford	None
Neil Gibson (representing the Royal College of Paediatric and Child Health) Consultant in paediatric respiratory medicine, Royal Hospital for Sick Children, Glasgow	None
Jill Gordon Consultant community paediatrician, Suffolk Primary Health Care Trust	None
Imelda Hughes Consultant paediatric neurologist, Royal Manchester Children's Hospital	None
Renee McCulloch Consultant in paediatric palliative medicine, NBK Hospital, Ministry of Health, State of Kuwait Senior clinical lecturer, Great Ormond Street Hospital and Institute of Child Health, London	None
Rob Ross Russell Consultant in paediatric respiratory medicine and paediatric intensive care, Cambridge University Hospitals	None
Anita Simonds Consultant in respiratory medicine, Royal Brompton and Harefield Hospital, London	Educational grant from ResMed Co; Steering Committee member for the Serve-HF trial in adults with heart failure and central sleep apnoea
Sian Haigh Lay member and parent	None

Guideline Endorser(s)

Royal College of Paediatrics and Child Health - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the British Thoracic Society Web site .

Availability of Companion Documents

The following is available:

- British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. Data supplement. (e-appendices). Available from the Thorax Journal Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on August 6, 2012. The information was verified by the guideline developer on August 17, 2012. This summary was updated by ECRI Institute on June 2, 2016 following the U.S. Food and Drug Administration advisory on Opioid pain medicines.

Copyright Statement

Disclaimer

NGC Disclaimer